MONGOLIAN IDIOCY

OCCURRENCE IN OF TWINS—REPORT OF TWO CASES

By HENRY DIETRICH * AND HUGH K. BERKLEY

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Mongolism in successive members of the same family is very rare.

The physical signs of mongolism show but slight variations in individual cases.

Mongolism may be complicated by rickets, hypothyroidism, adenoid growth, nad congenital heart lesions.

There is no successful treatment for the uncomplicated disease. Intelligent treatment may improve complicating endocrine gland disturbances, but has no effect upon mongolism itself.

DISCUSSION *by William M. Happ, Los Angeles; W. P. Lucas, San Francisco.*

MONGOLIAN idiocy is distinguished from other forms of idiocy by the fact that certain definite structural peculiarities of the body are associated with serious impairment of the mental functions. The condition was first described by Langdon Downs, an English physician, in 1866. At present it is estimated that from 3 to 5 per cent of all cases of congenital imbecility are of the mongol type. Several authors are of the opinion that the disease, of late years, is on the increase. This may be a true statement of fact; on the other hand, it is also quite possible that the apparent increase is due to its more frequent recognition by the medical profession. Be that as it may, its common occurrence, together with the fact that both the laity and physicians often fail to recognize mongolism, justify a short resumé of the subject.

The etiology of this peculiar disturbance in development is still obscure. H. H. Goddard collected 294 cases of mongolian idiocy in children, of whom 51 per cent were the last born of more than one child. Leeper collected 176 cases, and reports that no less than 51 per cent of these children were the last born of large families. These reports, together with others, led to the statement that mongolism is a disease due to exhaustion of the reproductive functions. Thursfield, in 1921, reviewed 42 cases. He could find no corroboration of the statement that a mongol is apt to be the last born child of a family in which the mother was near the end of her child-bearing period. He also states that syphilis, tuberculosis, and evidence of ill-health during pregnancy were not noted in these cases.

Stoltzner reported that the mothers of the patients in his series showed signs of hypothyroidism. Dollinger, in a study of twenty-five cases, could not confirm these findings.

Sajous, Sr., called attention to the relationship of insufficient functioning of the thymus gland to certain types of low mental development, particularly mongolian idiocy. One after another, and at times collectively, disturbances of the endocrine glands have been called upon to help explain this complex condition. The thyroid, especially, has been suspected of being at play, because of some resemblance between the physical characteristics of mongolism and those found in myxoedema.

T. Halbertsma advanced the theory that mongolism is the result of defects inherent in the germ plasm. He says: "When we have mongolism in one of twins, we have a pregnancy of the two-egg type; each twin has its own chorion and amnion. In such cases, two ova are simultaneously fertilized and develop independently. In contra-distinction, twins resulting from one-egg pregnancies are practically identical and inherit the same disease present in the germ cell. If mongolism were not germinal, but acquired during inter-uterine life, the occurrence of mongolism in one of twins would be an anomaly."

Mongolism in successive members of the same family is very rare. Vander Sheer, in reporting two such cases, one with two mongols and the other with three, states that he has been able to find on record only eleven families in which more than one child was a mongolian idiot. Since that time four more cases have been reported, making a total of fifteen. We have among our records another case to add to this list. The older of the children, a boy, is now 9 years old. His sister died at the age of 3 years from bronchopneumonia. She also had a very severe congenital heart lesion. Both were pronounced mongolian idiots.

Babonneaux and Villette reported one instance of four cases in one family. Halbertsma, in reviewing the situation, up to 1923 found only fifteen cases of mongolism in one of twins, and only two of mongolism in both twins.

Mongols rarely attain adult life. We see a great many instances of mongolism in infancy and early childhood, but the majority are carried off by intercurrent disease, principally of the respiratory or gastro-intestinal tracts, before they reach the age of 10 years. William N. Berkeley cites the following remarkable case, reported by Dr. Pogue, of a mongolian idiot girl who grew up and married, had one miscarriage, and subsequently gave birth to a child, at full term, who was not a mongolian idiot.

The physical signs of mongolism show but slight variations in individual cases. The following signs are found in practically every case of mongolism: Slanting eyes, the inner canthus being lower than the outer, narrow lid apertures; brachycephaly, epicanthus, small, saddle-shaped nose, external ears atavistically malformed; gaping mouth, frequently with protruding tongue, prominent abdomen, diastasis of the recti, umbilical hernia, shortened and incurved little finger, due to hypoplasia of the distal phalanx; delayed closure of the fontanelle, delayed and irregular teething, flacidity of muscles and joints, lack of resistance to intercurrent diseases and definitely impaired mentality.

Many of the children are not able to sit up until 18 months to 2 years of age, and do not walk or talk util 3 or 4 years old. They rarely learn to speak correctly, often maintain uncleanly habits,

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and usually do not attain a mental development beyond that of a 5 to 6-year-old child.

The demeanor of the mongolians is very characteristic. During the first year or two they are apathetic, often difficult to feed, and the mother remarks about her exceptionally good child. After that period, they become aggressive, restless, with a tendency to gesticulation and imitation. The disposition is lively and happy, and this change is then regarded by the layman as an evidence of progress; but unfortunately it rarely leads to anything beyond a clownish, imitative child. A love for music is noticeable in many of these children. They are not capable of school training. It is the duty of the state to furnish a proper means for educating these children, but up to the present time very inadequate provision has been made.

Fritz Talbot reports a series of observations on the growth of untreated mongolian idiots, ten in number, ranging from 4 months to 10 years of age. "There are several factors of growth in common between the cretin and the mongolian idiot. The arms, legs, and feet of both conditions tend to be shorter than the normal—more so in the former than in the latter. The greatest difference in their physical measurements is noted in the circumference of the head; that of the mongolian idiot is less than normal, while that of the cretin falls within normal limits."

Mongolism may be complicated by rickets, hypothyroidism, adenoid growth, and congenital heart lesions. The marked susceptibility of these children to respiratory infection and gastro-intestinal diseases must be emphasized.

There is no successful treatment for the uncomplicated disease. Intelligent treatment may improve complicating endocrine gland disturbances, but has no effect upon mongolism itself.

CASE I—Male child, age 8 weeks, one of twins, first born. Mother 26, father 28 years of age. No miscarriages. No illness on part of mother during pregnancy. Birth weight, 5 pounds. Has had a wet nurse. Now weighs 7 pounds 5 ounces. Parents state child makes peculiar noises when breathing. Is much more quiet than its twin, and seems to have no strength in back of neck. Sleeps a great deal. Is constipated. Physical examination shows a child with slanting eyes, marked epicanthus, short, wide nose, marked umbilical hernia, incurved little finger and supernumerary thumb on left hand. Very wide-open anterior fontanelle, post fontanelle 2½ x 2½ cm., and sagital sutures separated for 1 cm. Flaccidity of muscles and subcutaneous tissue. Ears small, deformed, and ear canal is very small. Child holds tongue between lips continually. Bilateral hydrocele. Heart shows no evidence of congenital malformation.

Diagnosis—Mongolian idiocy.

The twin was a male child, 8 weeks of age. Birth weight 5 pounds 6 ounces. Now weighs 8 pounds 3 ounces. Physical examination reveals nothing abnormal, except that post-fontanelle is still open.

CASE II—Female child, 5 months old, one of twins. Ninth child. Mother 37 years, father 40. No miscarriages. Father and mother in good health. Birth weight 5¾ pounds. This child was born first, was very cyanotic after birth. Could not feed at the breast during the first week, but since that time is able to suckle the breast, but with very poor result, as she only weighs 6 pounds at present. Dr. Homer, who referred the case to us, reports a double placenta. Physical examination shows an atrophic child with slanting eyes, epicanthus, fine, reddish hair. Large anterior fontanelle and post-fontanelle still open 1 x 2 cm. Marked hypotonicity, incurved little finger, narrow chest, deformed ears, tongue is almost constantly protruded. A congenital heart lesion was present.

Diagnosis—Mongolism.

The other twin, also female, we have not seen. However, Dr. Homer of Ventura reports that she weighed 8½ pounds at birth, and has steadily thrived and developed. Photographs he has kindly sent us show a well-developed child, with no suggestion of mongolism.

DISCUSSION

WILLIAM M. HAPP, M. D. (523 West Sixth Street, Los Angeles)—The occurrence of mongolian idiocy in one of twins as reported by Drs. Dietrich and Berkley is extremely interesting, and, it seems to me, lends added weight to the view that the condition is due to defective germ plasm, and is, therefore, a developmental defect. As the authors state, there is no evidence that the thyroid or other endocrine glands are responsible for the condition. I think that the far too common practice of treating mongolian idiocy with thyroid or polyglandular therapy is withut scientific basis. Personally, I have never seen a case show any improvement under such treatment beyond the normal improvement these children all show.

W. P. LUCAS, M. D. (490 Post Street, San Francisco)—The paper of Drs. Dietrich and Berkley adds two additional cases to a rapidly growing list of mongolian idiocy in one of twins. We have observed no cases of mongolian idiocy in one of twins at the University of California Hospital, although the condition itself is one which is very frequently seen.

Physiological and pathological studies have so far failed to give any clues as to the etiology of mongolian idiocy, and there exists no therapy which has the slightest influence. Talbot has shown that in some cases it is possible to produce a brief initial improvement by the administration of thyroid extract. This is unquestionably due to the associated glandular involvement in this condition. The improvement stops just as soon as the thyroid component of the clinical picture has been repaired, and one is then again confronted by a stationary condition which is resistant to all treatment.

Prevention, rather than treatment, would seem to be the direction from which help will eventually come. And the accumulation of statistical evidence, to which Dietrich and Berkley have added two important cases, is one of the most promising methods of approach.

"After the death of Dr. Conyers Middleton (whom I have had occasion to speak of before, as the author of the attack on the dignity of physic, which was so warmly and triumphantly repelled by Dr. Mead), his widow called upon Dr. Heberden with a MS. treatise of her late husband, about the publication of which she was desirous of consulting him. The religion of Dr. Middleton had always been justly suspected, and it was quite certain that his philosophy had never taught him candour. Dr. Heberden having perused the MS., which was on the inefficacy of prayer, told the lady that though the work might be deemed worthy of the learning of her departed husband, its tendency was by no means creditable to his principles, and would be injurious to his memory; but as the matter pressed, he would ascertain what a publisher might be disposed to give for the copyright. This he accordingly did; and having found that £150 might be procured, he himself paid the widow £200, and consigned the MS. to the flames."—"The Gold-Headed Cane."

Dr. Marriott of St. Louis recently visited Southern California and gave a number of lectures and clinics at the Scripps Metabolic Clinic, and before the Southwestern Pediatric Society, and the San Diego Medical Society.

These conferences proved so stimulating and encouraging that, under authority of the San Diego Medical Society, a committee is already engaged in arranging for a series of lectures for 1927. David H. Higbee, Watts Building, San Diego, is chairman of the committee.